Golestan University of Medical Sciences
Journal of Clinical and Basic Research
Online ISSN: 2538-3736

Case Report

Acute Pancreatitis in the Setting of Vaso-Occlusive Sickle Cell Crisis: A Rare Differential Diagnosis

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ReceivedMay22, 2022 **Received in revised form**June3,2022 **Accepted**July3,2022

ABSTRACT

Background: Sickle cell disease (SCD) is a hemoglobinopathy characterized by chronic hemolytic anemia and vaso-occlusive painful crisis. The vascular occlusion in SCD is a complex process that is responsible for most clinical manifestations of the disease. Abdominal pain is an important component of vaso-occlusive painful crisis and may mimic diseases, such as acute appendicitis and cholecystitis. Acute pancreatitis is rarely encountered as a cause of abdominal pain in patients with SCD. Gallstones and alcohol abuse account for the majority of the cases of acute pancreatitis. Pancreatic ischemia is an uncommon but established cause of pancreatitis associated with connective tissue diseases, vasculitis, and shock. In SCD, the deformed red blood cells resulting from deoxygenation during stress, infection or dehydration, cause vaso-occlusion, tissue ischemia, and infarction.

Case description: We present a case of SCD presenting as acute pancreatitis apparently due to microvascular occlusion and ischemic injury to the pancreas. The patient responded to conservative management.

Conclusion: Our case will help to always keep in mind and consider acute pancreatitis as a differential diagnosis in patients with SCD presenting with abdominal pain.

Keywords: Acute pancreatitis, Sickle cell disease, vaso-occlusive painful crisis

DOI: 10.29252/Jcbr.6.2.31

How to Cite: Sahu D, Nayak S, Mohanty N, panda B. Acute Pancreatitis in the Setting of Vaso-Occlusive Sickle Cell Crisis: A Rare Differential Diagnosis.Journal of Clinical and Basic Research. 2022; 6 (2) :31-35

Introduction

Sickle cell disease (SCD) is an autosomal recessive disease and one of the most common hemoglobinopathies, accompanied with production of hemoglobin S (Hb S) due to a point mutation in the beta globin gene. It is characterized by chronic hemolytic anemia and repeated episodes of vaso-occlusive painful crisis (1,2). Deoxygenated HbS causes distortion of red blood cells (RBCs) when the oxygen saturation is lowered in conditions such as stress, infection, or dehydration. The deformed RBCs cause vaso-occlusion, tissue ischemia, and infarction. Virtually all organs in the body are affected by vaso-occlusion. Acute pancreatitis has been described as a very rare manifestation of vaso-occlusive painful crisis (3,4). Gallstones and alcohol abuse are the major causes of acute pancreatitis (5). Pancreatic ischemia is an uncommon but established cause of pancreatitis associated with connective tissue diseases, vasculitis, and shock (6). We present a case of acute pancreatitis as a rare manifestation of vaso-occlusive painful crisis in a patient with SCD, presumably due to microvessel occlusion and ischemic injury to the pancreas.

Case Presentation

A 20-year-old male patient with a history of heterozygous SCD (Hb SS)-beta thalassemia since 2008 presented to the emergency department (ED) with diffuse abdominal pain and back pain for 7 days, which worsened over last 2 days. He had a history of multiple past hospitalizations for vaso-occlusive crises. He had no history of alcohol consumption. He described the pain as sharp, diffuse, and more severe in the epigastrium, which worsened with movement and did not relieve by oral analgesics. In ED, his pulse rate was 120/minute and low volume, blood pressure was 90/60 mmHg, respiratory rate of 24/minute, oxygen saturation of 98% in room air, and temperature of 97.8°F. On physical examination, he had a toxic appearance with scleral icterus. The patient also had diffuse abdominal tenderness, more at epigastrium, but with no distension. There was no guarding, no rigidity or rebound tenderness. Bowel sounds were present. Rest of the systemic examinations were not significant.Immediate intravenous access was obtained. Fluid resuscitation was initiated. All baseline laboratory tests as well as blood and urine cultures were taken. Broad spectrum antibiotics (piperacillin/tazobactam) were initiated due to the undifferentiated nature of the hypotension at that time.

Laboratory investigations revealed hemoglobin of 8.1 g/dl and white blood cell count of 38.01×109/l. Reticulocyte count was increased to 3.1%, and the HbS level was 51.1%. The basic metabolic panel revealed serum urea of 18 mg/dl, serum creatinine of 0.6 mg/dl, blood glucose of 120 mg/dl, total bilirubin of 7.2 mg/dl, indirect bilirubin of 4.5 mg/dl, aspartate transaminase of 42 U/l, alanine transaminase of 138 U/l, alkaline phosphatase of 261 U/l, lactate dehydrogenase (LDH) of 701 U/l, serum amylase of 108 U/l, and serum lipase of 322 U/l. The lab parameters suggested hemolysis as evidenced by unconjugated hyperbilirubinemia and increased LDH. Ultrasonography of the abdomen revealed hepatosplenomegaly with multiple echogenic foci in splenic parenchyma (Gamna-Gandy bodies), mild bulky pancreas with a normal biliary tree, and gallstones without evidence of obstruction. A contrast computed tomography (CT) scan of the abdomen showed mild hepatosplenomegaly with splenic infarct and peri-splenic collection, mild bulky pancreas with mild peripancreatic stranding (acute pancreatitis), and CT severity index score of 2(Figure1). Acute pancreatitis was diagnosed, and the patient was on nil per oral, intravenous fluid, and antibiotics (piperacillin/tazobactam). Over the next 7 days, his pain resolved and the lipase and amylase levels decreased to 34 and 44 U/l, respectively. Moreover, LDH and reticulocyte count decreased to 412 U/l and 2.4%, respectively. Oral intake was resumed, and the patient was discharged.

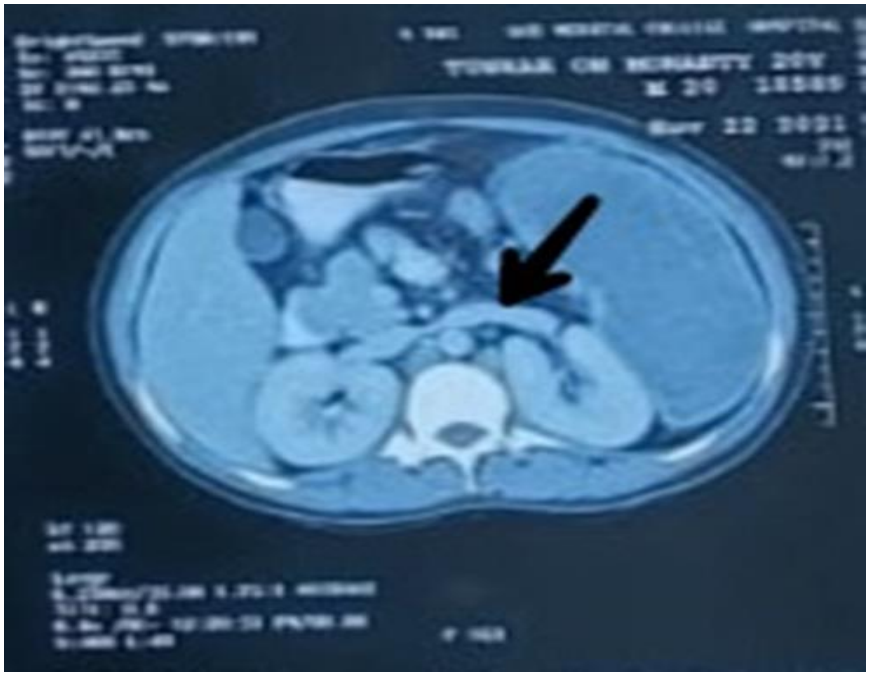

Figure 1. CT of the abdomen showing mild bulky pancreas with mild peripancreatic stranding (black arrow), consistent with acute pancreatitis

Discussion

Abdominal pain is a common manifestation of sickle cell painful crises and may result from ischemic infarcts of the mesentery and abdominal viscera due to microvascular occlusion (7). Almost all abdominal organs can be affected by SCD, secondary to sickling, hypercoagulability, capillary engorgement, and stasis in the vasa vasorum of larger vessels (8). In majority of cases, no specific precipitating cause is identified, and symptoms resolve with supportive measures. The clinical features of abdominal pain due to SCD vaso-occlusive crisis may be indistinguishable from those of acute intra-abdominal disease processes, such as acute cholecystitis, hepatic infarction, splenic sequestration, renal papillary necrosis, urinary tract infection, peptic ulcer disease, ischemic bowel, and appendicitis (1,7). Despite a high incidence of gallstones, acute pancreatitis is rarely included as a cause of abdominal pain in patients with SCD, who are at risk for acute pancreatitis both from biliary obstruction and potentially from microvessel occlusion with resultant ischemia, activation of pancreatic enzymes, and pancreatic injury (3,4,9). Acute ischemic pancreatitis is a very rare complication of vaso-occlusive crisis.

The clinical features are indistinguishable from other causes of acute abdomen and represent a diagnostic challenge. Provisional diagnosis of acute pancreatitis is based on the presence of abdominal pain and biochemical evidence of pancreatic injury (10). Serum amylase and lipase levels have been considered as the biochemical standard for diagnosing acute pancreatitis. Although serum amylase may be elevated in several other conditions, such as perforated peptic ulcer or ischemic bowel, the increase is rarely more than three times the normal level. Serum lipase has a higher sensitivity and specificity and therefore has more reliability than serum amylase for the diagnosis of acute pancreatitis (11). In addition, CT is a reliable and non-invasive imaging modality to evaluate the pancreas. Contrast-enhanced CT is the diagnostic technique of choice because of its ability to demonstrate the presence and extent of necrosis (12). There was no evidence of drug, alcohol, trauma, toxin, or an obstructive etiology, which suggests that

pancreatitis was likely due to ischemic etiology from sickling (7). Although splenic infarct can cause abdominal pain, the elevated serum amylase and lipase levels along with the radiological findings in our case suggested acute pancreatitis as the cause of abdominal pain.

Similar to any other case of acute pancreatitis, treatment is predominantly conservative and includes intravenous hydration, pain control, and electrolyte replacement. Exchange transfusion may be the therapy of choice in patients with acute multi-organ failure (13).

Conclusion

Acute pancreatitis due to ischemic in patients with SCD may be important to consider in the differential diagnosis of abdominal pain. Abdominal pain in patient with SCD presents a diagnostic challenge. While pain from a vaso-occlusive crisis is a common etiology, acute pancreatitis should also be considered due to a vaso-occlusive episode as suggested by our case report. Deviation from the usual pattern of pain and/or failure to respond to hydration and supportive care should perhaps increase the clinician's suspicion of this entity.

Declarations

Funding

Not Applicable

Ethics approvals and consent to participate

The patient was informed from the research condition and the informed consent letter was taken.

Conflict of interest

The authors declare that there is no conflict of interest regarding publication of this article.

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